Gastroenterology and Hepatology From Bed to Bench.

CASE REPORT

A patient with cystic lymphangioma in pancreas

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ABSTRACT

Cystic lymphangioma of the pancreas is extremely rare, accounting for less than 1% of pancreatic tumors. Though congenital, it can affect all age groups. Cystic lympangioma occurs more frequently in females. Patients usually present with epigastric pain and an associated palpable epigastric mass. Complete excision is curative, even though, depending on the tumor location, surgery may be simple or involve extensive pancreatic resection and anastomoses. In this case report, we discuss a 63-year old patient who presented with epigastric pain and on investigation was found to have pancreatic head cystic lymphangioma. At surgery the tumor was completely excised, with preservation of pancreatic duct. Histology and immunohistochemistry confirmed cystic lymphangioma of the pancreas. This case highlights that a diagnosis of cystic lymphangioma of the pancreas should be taken into consideration as a differential diagnosis of pancreatic cystic lesions.

Keywords: Lymphangioma, Pancreas, CT scan.

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Introduction

Cystic lymphangioma arise as a result of a lymphatic vessels developmental failure. This condition was described for the first time by Koch in 1913 (1). It probably occurs as a result of a congenital malformation of the lymphatics resulting in obstruction of the local lymph flow and the development of lymphangiectasia (2). Pathologically, it exhibits protein and eosinophilic exudates, within a swollen cystic space, in a thin layer surrounded by the endothelial cells (3). Cystic lymphangiomas are usually benign, but can be locally invasive (4). Cystic lymphangioma can occur in any location where normal lymphatic ducts are found (3). Cystic lymphangioma are most commonly found in the neck, the axilla and the mediastinum in children (2). It is rare in adult population (5, 6). The disease is reported more often in females, with a similar incidence across all age groups (3).

Cystic lymphangioma inside the abdominal cavity is very rare, occurring most frequently in the mesenteric region, and less frequently in the mesocolic, greater omental and retroperitoneal regions, small intestinal and gastric walls, and peritoneum (3). A variety of other sites have been described including the mediastinum, pleura, pericardium, groin, bones and abdomen. In the abdomen, the mesentery and retroperitoneum are the most common sites;

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pancreatic cystic lymphangioma constitutes 1% of all lymphangioma (7, 8). Pancreatic cystic lymphangiomas may clinically mimic pancreatic carcinoma and should be considered as a differential diagnosis in any patient found to have an abdominal mass (9). To our knowledge, we are unaware of a reported case of pancreatic cystic lymphangioma from Iran. We present the rare case of an adult with lymphangioma of the head of the pancreas and review the literature.

Case Report

A 63-year-old female presented with a year history of vague abdominal pain, anorexia and malaise. The pain was colicky in nature, localized to the epigastric region, with radiation to the interscapular region of the back. Pain was reduced by changes in sitting position and increased by eating. There was no history of alcohol consumption or abdominal trauma. Routine investigations revealed a thrombocytosis (PLT>1000000 cells UL^{-1}) and raised urea (500 mgr/daily). Aspirin, (80 mgr/daily) was prescribed for essential thrombocytosis. An abdominal ultrasound was suggestive of a cystic lesion at the pancreatic head.

An abdominal computerized tomography (CT) scan with intra venous and oral contrast was performed and a cyst with a maximum diameter of 11.8 cm was seen (Fig 1); the mass had not invaded any adjacent organs. Moreover there was no evidence of lymphadenopathy. The CA 19-9 tumor marker was within the normal range, less likely making the diagnosis of malignancy. The patient was referred to the Shohadaye-Tajrish hospital for surgical assessments in November 2010. The patient had a past surgical history of an excisional biopsy due to lymph nodes enlargement in right axillary region and a history of psychological disorders (moderate depression) was also noted (treated with Perphenazine 2 mg thrice daily, Clonazepam 1mg once daily and Nortrilen 25mg once daily). There was no history of ischemic heart disease, diabetic mellitus, and hypertension. On physical examination, the patient had pleuritic face, vital signs were normal and auscultation of the heart and lungs was also normal.

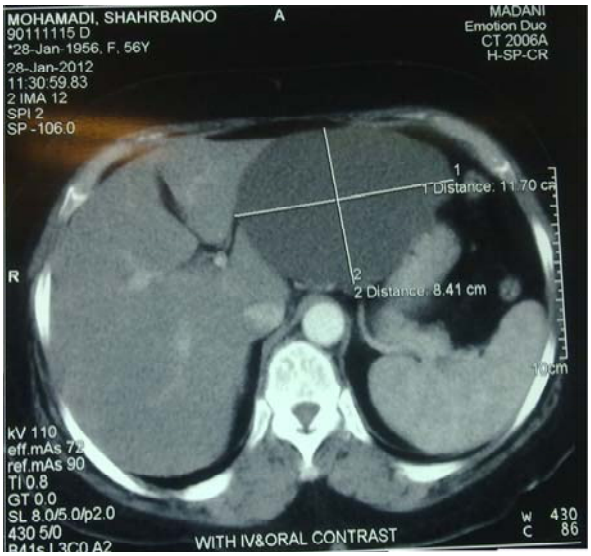

Figure 1. Abdominal computerized tomography (CT) scan with intra venous and oral contrast showing a 11.7×8.41cm cystic mass of pancreas

The abdomen was not distended, with normal umbilicus and superficial veins. There were no signs of portal hypertension or previous surgery. Mild tenderness upon deep palpation of the epigastrium was noted, without rebound tenderness or rigidity. Following diagnostic imaging surgical exploration at laparotomy revealed a 12 cm single cystic mass at the head of the pancreas, not attached to any adjacent organs and without connection with main pancreatic duct (Fig 2). Considering the risk of malignancy and seeding, we avoided biopsy and puncture. The patient underwent cystectomy procedure and the main pancreatic duct was preserved, with a primary diagnosis of pancreatic pseudocyst. No other pathology was found within the abdomen and pelvic.

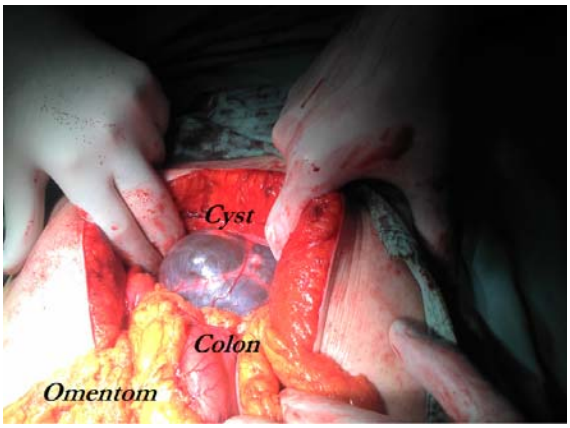

Figure 2. Laparotomy exploration showing a large cyst and location

On gross examination, the tumor, measuring 115×85mm, had a nodular, gray-blue surface and was indented with normal head of pancreatic tissue (Fig 3).

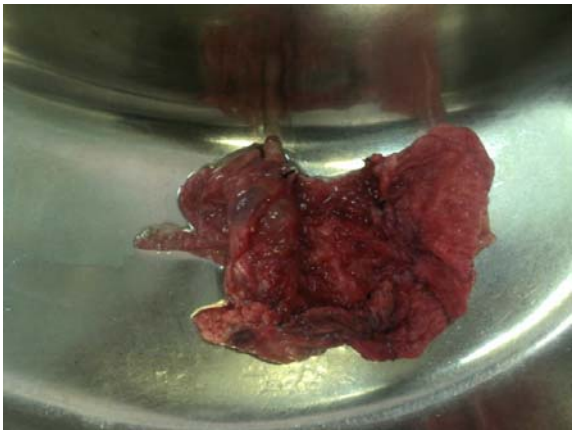

Figure 3. Nodular, gray-blue surface tumor measuring 115×85mm on gross examination after removal cyst

On gross sectioning, the cyst had a honeycomb appearance with single space filled with murky haemorrhagic yellowish fluid. Histology sections showed cystic lesions of variable size, separated by fibroconnective septa, containing irregular smooth muscle fascicles, adipocytes and mature lymphocytes (Fig 4).

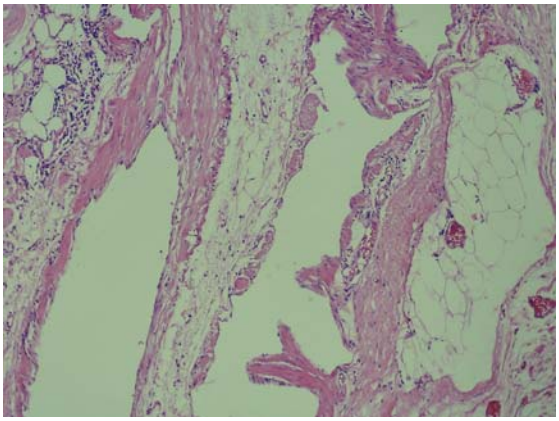

Figure 4. Histological findings showing cystic wall contains irregular smooth muscle fascicles, adipocystes and mature lymphocytes

These cystic spaces were lined by flattened and slightly elevated endothelial cells (Fig 5). No cell atypia was found.

Immunohistochemistry (IHC) labeling for endothelial markers, factor VIII-R antigen and CD 31 were positive while CD 34 was negative. In keeping with a diagnosis of cystic lymphangioma, the patient was discharged 3 days after surgery, following an uneventful postoperative recovery. Her abdominal pain resolved and investigations including normalized serum urea and creatinine. She is still doing well with a good quality of life and no complaints.

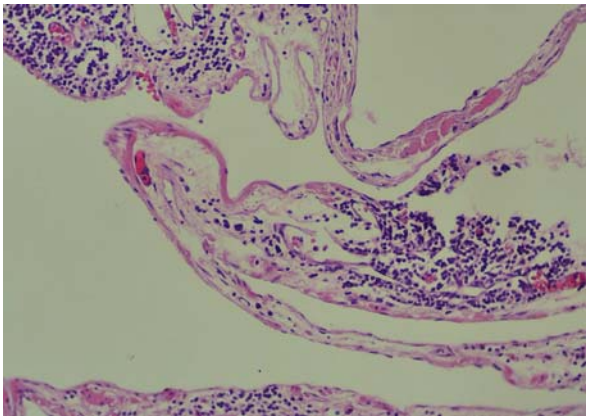

Figure 5. Histological findings showed that the cystic spaces are lined by flattened endothelial cells.

Discussion

Intraperitoneal lymphangiomas occur most frequently in the in the mesenteric region and less frequently in the mesocolic, greater omental, and retroperitoneal regions,the small intestinal and gastric walls, and the peritoneum (3). Cyctic lymphangioma rarely occurs in pancreas, accounting for less than 1% of lymphangiomas[6]. Several factors are assumed to cause cystic lymphangioma. These include lymphatic abnormalities resulting in children from sequestration of lymphatic tissues during prenatal development and in adults from blockage of lymphatic ducts due to abdominal trauma, operation, radiological treatment, or infection (2). lymphangioma are classified into three types: simple capillary, cavernous and cystic (6, 10). Cystic lymphangiomas of the pancreas are composed of dilated cystic spaces lined by flattened endothelium containing abundant lymphoid tissue and smooth muscle in the wall of the cyst (7). As a result, reports in the literature are limited to case reports with very few case series. Most reports in the literature describe cystic lesions in the body and tail of the pancreas as these are the most common regions involved (11). There have only been 16 previously reported cases of this tumor arising from the head of the pancreas (12, 13). Pancreatic pseudo cyst, cyst adenoma, congenital cyst, rare pancreatic ductal carcinoma (cystic carcinoma of the Wirsung's Duct), and congenital neoplasms are among th differential diagnosis (1, 10). To ensure distinction between cystic lymphangioma and other conditions, diagnostic tools such as plain abdominal films, abdominal CT, and magnetic resonance imaging (MRI) may be helpful. Plain abdominal X-rays may reveal dislocation or blockage of the intestines, and CT and MRI could help determine the pre-operative location of the mass, and facilitate determining the possibility of communication between the mass and an adjacent structure, the size of the mass and likely complications of surgery. However, these tests do not guarantee a completely accurate diagnosis. It is still considered a difficult task to accurately diagnose pancreatic cystic lymphangioma prior to a surgical procedure (2, 14, 15). This patient had no previous history of pancreatitis. Therefore, we could safely rule out the possibility of this lesion being a pancreatic pseudo cyst. We were looking for signs of invasive malignancy, namely ascites, peritoneal lesions, lymph nodes and involvement of the adjacent peripancreatic organs, all of which were ruled out during this procedure. Clinical symptoms of cystic lymphangioma found inside the abdominal cavity, if present, vary from no symptoms to a variety of symptoms depending on the location and size of the mass. More acute cases are reported in children whereas more chronic development is common among adults (3). Features of pancreatic cystic lymphangioma normally include abdominal pain and the presence of a mass confirmed by physical examination. Patients usually present with abdominal pain (11). Pancreatitis, weight loss, and laboratory abnormalities are not usual disease manifestations (16). On CT, the tumor is a well-circumscribed, encapsulated, water-isodense, polycystic tumor with thin septa, similar in appearance to cystadenomas, which occur far more frequently (4, 16).

Immunohistochemistry can be used to increase the accuracy of diagnosis. Since the endothelial cells of the outer membrane of pancreatic cystic lymphangiomas do not normally undergo epithelial differentiation, staining materials, such as (Periodic Acid-Schiff) PAS stain, Mucicarmine, Alcian blue, and anti-cytokeratin are unsuitable. However, staining with IHC materials like Factor VIII-R Ag, CD31, CD34, and D2-40 is helpful. Diagnosis of the disease,

therefore, involves the use of such IHC (2, 17). The final diagnosis is with histopathology and IHC for endothelial cell markers such as factor VIII-R antigen and CD31 positively and CD34 negatively, as seen in our patient (16). If the infiltrated organs are excisable, they must also be removed along with the mass itself with the aim of treatment being a benign excised surface on microscopic examination (14, 15). A complete surgical excision is curative for lymphangioma, with incomplete excision being the only reason for recurrent disease (6). Depending on the tumor location and size, complete excision may involve a simple excision of the mass or may require pancreatic resections, such as a Whipple procedure or distal pancreatectomy with anastomosis (18). We therefore report this rare case in the hope that although extremely rare, cystic lymphangioma of the pancreas should be taken into consideration as a differential diagnosis of pancreatic cystic or retroperitoneal lesions, especially in women.

Cystic lymphangioma of the head of the pancreas is very rare but lymphangioma of the pancreas should be taken into consideration as a differential diagnosis of a pancreatic cystic lesion. Treatment is complete excision. Incomplete excision is a reason for recurrent disease.

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